Isolated Cutaneous Langerhans Cell Histiocytosis in a Three Months Old Infant- A Case Report

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Introduction

• Cutaneous Langerhans Cell Histiocytosis (LCH) is a rare disorder which typically presents in children under two years of age with a variable clinical course ranging from spontaneous resolution to fatal outcome.

• LCH in infants can be similar in presentation to other pediatric rashes such as atopic dermatitis, seborrheic dermatitis, arthropod assault and diaper dermatitis. Misdiagnoses can result in delayed diagnosis and ineffective treatment due to low awareness and understanding of LCH.

• We present the case of a three months old infant promptly diagnosed with LCH following biopsy of the lesion after unsuccessful treatment for atopic dermatitis and arthropod assault.

• Given the rarity of the condition and variable clinical presentations, there is no consensus treatment guideline of LCH. This case report outlines the initial clinical work-up and treatment of an infant with isolated cutaneous LCH and subsequent response to therapy.

Case Report

A three-month-old Hispanic female presented to dermatological consultation with generalized pruritic erythematous papules, pustules and nodules on the scalp, trunk, and bilateral upper and lower extremities of two months’ duration (Figures 1 & 2). She was empirically treated by her pediatrician with scabies therapy followed by dermatological consultation with generalized pruritic erythematous papules, pustules and dermatitis on the upper and lower extremities of the infant.

Apart from the lesions on her skin, the rest of the physical exam was unremarkable. She did not appear to be in pain when held or picked up. Her weight and growth were within normal percentiles for her age group. Her mucous membranes were uninvolved. There was no cervical, axillary, or inguinal lymphadenopathy. There was no visceral enlargement on exam.

Laboratory and Diagnostic Findings: A papule on the right hip of the infant was biopsied and revealed superficial and deep perivascular, perineural and mixed inflammatory cell infiltrate in the dermis and extending to the subcutaneous fat containing lymphocytes, histiocytes and numerous eosinophils. The diagnosis of Langerhans cell histiocytosis (LCH) was made based on morphologic criteria and confirmed by positivity of abnormal Langerhans cells by anti-CD1a immunostain (Figure 2a & 2b).

The baby was referred to a pediatric oncologist for further evaluation and treatment. Chest x-rays were unremarkable. Blood count, serum urea, creatinine, bilirubin, liver enzymes, and urine osmolality were within normal limits. Total skeleton radiograph and scintigraphy failed to show any bone lesions. Workup for diabetes insipidus was negative. It was concluded that the baby had a case of isolated cutaneous LCH.

Intervention: Treatment was initiated with oral low dose methotrexate (20mg/m2) for a planned duration of six months with disease evaluation every two weeks for routine labs and disease monitoring. Chemotherapy regimen consisting of vinblastine and an oral steroid was decided as the alternative treatment should disease progress to multisystem LCH.

Response to Treatment: After two months of methotrexate therapy, the patient significantly improved with a decrease in the overall number of lesions. No adverse effect to treatment has been noted and baby is still undergoing therapy.

Discussion

• LCH is a rare disease of idiopathic monoclonal proliferation of abnormal Langerhans cells and cytokine overproduction resulting in inflammation, infiltration and destruction of many tissues in the body.

• There is poor understanding of the true incidence and burden of LCH due to the rare nature of the disease and its diverse clinical presentations.1,2

• Clinical features of LCH can range from localized, single-organ lesions to multifocal, multi-organ involvement.

• LCH confined to the skin as presented in this report is rare and accounts for only about 5% of LCH cases.

• LCH can result in spontaneous regression, re-occurrence or progress to rapid deterioration, and even death.3-4 Isolated cutaneous LCH tends to regress spontaneously, but progression to MS-LCH is also common.4

• There is no consensus management guideline for LCH and the rarity of the condition complicates proper research on the most effective treatment.

• For LCH recalcitrant to topical steroid therapy where there is involvement of an extensive area, systemic therapy with oral steroids with or without vinblastine or oral low dose methotrexate can be used. It should however be noted that the level of evidence with this therapy is low.5

Conclusions

• LCH should be considered in the differential diagnosis of disseminated papulo-pustular lesions in infants; especially those recalcitrant to treatment.

• The unpredictable nature of the disease underscores the need for close follow up and careful observation in order to identify in time progression to potentially fatal systemic disease.

References

5. Up, and...